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## **Long-gap esophageal atresia: management, most frequent complications, and expert recommendations – review of literature**

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**Abstract**

**Introduction and aim of the study.** Long-gap esophageal atresia (LGEA) is a congenital anomaly in which the gap between both ends of the esophagus exceeds three intervertebral spaces and is an esophageal atresia without air in the abdomen. The defect is both therapeutic and surgical challenge. This review aims at providing an overview of the most recent

literature on the effective methods for treatment of LGEA, and the most frequent complications and experts' recommendations on this subject.

**Material and methods.** The systematic review was based on available data collected using PubMed database and the Google Scholar web search engine.

**Analysis of the literature.** There is no consensus on the ideal technique for surgical treatment of LGEA. There are two possible approaches for opening the thorax – thoracotomy and thoracoscopy. The techniques stimulating esophageal elongation include external and internal traction techniques, magnetic compression anastomosis and intramural botulinum type A toxin injection. Replacement methods are a viable option when it is impossible to preserve the native esophagus. Decellularized matrices seem to be promising in developing an esophageal substitute. Regardless of the surgical approach a common complication of surgical treatment is anastomotic stenosis which requires further surgical interventions.

**Conclusion.** Elongation techniques are effective in approximation of the proximal and distal esophagus. The future lies with tissue engineering and inventing an off-the-shelf esophageal substitute. The centralization of treatment is recommended. After discharge from hospital interdisciplinary outpatient assessment and care is required. Further prospective studies are needed to determine the optimal mode of treatment and prevent complications associated with LGEA.

**Keywords:** esophageal atresia, long gap, surgical management, delayed primary anastomosis, anastomotic strictures

## **Introduction**

Long-gap esophageal atresia (LGEA) is a congenital anomaly in which the gap between both ends of the esophagus exceeds three intervertebral spaces or is too long for early primary repair [1–3]. The European Reference Network for Rare Inherited Congenital Anomalies (ERNICA) defines LGEA also as any esophageal atresia without air in the abdomen[4]. The incidence of esophageal atresia (EA) is estimated at 1 per 2500-3500 live births, with a slight male predominance [1,5,6]. In about 6-7% it is a pure EA (Gross type A) without a fistula between the trachea and the distal esophagus[2,7,8].

LGEA may be an isolated defect or coexist with other malformations [1,6,7,9]. The most common are cardio-vascular anomalies. In some patients with EA chromosomal abnormalities, especially trisomy 18, are found, too. EA is also a component of the

VACTERL (Vertebral, Anorectal, Cardiovascular, Tracheo-Esophageal, Renal, and Limb anomalies) association and CHARGE (Coloboma, Heart defects, Atresia of the choanae, Retardation of development, Genital defects, and Ear anomalies and/or deafness) association[7,10].

It is possible to diagnose LGEA prenatally. The symptoms which suggest this congenital malformation are polyhydramnios, small stomach and blind proximal esophagus which can be visualized on ultrasound after 23 weeks of pregnancy [8,10]. After birth, in all cases with suspected EA, a nasogastric tube should be inserted and a thoracoabdominal X-ray should be performed as a diagnostic procedure [4]. A radiographic ‘gasless abdomen’ suggests pure EA[10].

The defect always requires surgical intervention. However, there is no consensus on the ideal technique [2,10]. Most researchers agree though, that the aim should be to preserve the patients’ own esophagus[1,4,11]. As regards the methods of opening the thorax there are two approaches – open (thoracotomy) and thoracoscopic [11]. Over the years several techniques stimulating esophageal elongation and enabling primary anastomosis have been developed and improved. These include external and internal traction techniques, magnetic compression anastomosis and intramural botulinum type A toxin injection[11–15]. In some cases, though, reconstructive surgeries, such as gastric or jejunal interposition need to be performed [1,3,4,10]. Nevertheless, future-oriented research focuses on developing an esophageal substitute using decellularized material with tissue engineering[16].

The most frequent, both early and long-term, complication of surgical treatment is anastomotic strictures (AS), which requires endoscopic dilation at the later stages of treatment [2,3,17]. Other frequent post-operative complications are anastomotic leakages, dysphagia, and gastro-esophageal reflux disease [1,3,18]. Patients with LGEA require treatment in experienced and well-equipped Centers of Expertise (CoE), as well as interdisciplinary follow-up programs due to the possible complications [4,16,19].

### **Aim**

This review aims at providing an overview of the most recent literature on the available, effective, and novel methods for treatment of LGEA, as well as an overview of the most frequent complications of surgical treatment and experts’ recommendations on this subject.

## **Material and methods**

This systematic review was based on the data collected using primarily the online databases, including PubMed database and the Google Scholar web search engine. The study was based on multiple meta-analyses, review articles, guidelines, research articles and case reports. We focused on the latest literature, over 70% of the articles being from years 2014 to 2023. Articles prior to 2014 were mainly used for analysis of the historical background.

## **Analysis of the literature**

LGEA is a congenital defect that is both a diagnostic and therapeutic challenge for pediatric surgeons and pediatricians. Even the definition of this malformation was difficult to standardize. In 2017 the experts of the International Network of Esophageal Atresia (INoEA) agreed that any esophageal atresia without air in the abdomen should be considered as a long-gap EA[19]. Two years later the American Pediatric Surgical Association (APSA) Outcomes and Evidence Based Practice Committee suggested that the definition of LGEA should not be the same as the definition of pure atresia. Moreover, they concluded that LGEA should not be defined by gap measurements, neither in centimeters, nor vertebral bodies [20]. Finally, in 2020 the experts of the ERNICA Consensus Conference on the Management of Patients with Long-Gap Esophageal Atresia accepted the LGEA definition suggested by the INoEA consensus[4]. Moreover, they proposed, in contrast to APSA Outcomes, that any esophageal atresia with a gap of three vertebral bodies should also be considered as a long gap[4].

Despite the variations in experts opinion regarding the definition, there is at least the consistency that this congenital defect always requires surgical intervention in the neonatal period or infancy. Management depends on the distance between the proximal and distal esophagus, and their condition[10,21]. Moreover, not only surgeon experience and skills, but also individual preferences affect the choice of a particular surgical approach [20]. Most researchers agree that the aim should be to preserve the patients' own esophagus [1,4,11]. Despite the enormous progress in tissue engineering there is still no ideal method for replacing a native esophagus[21].

Many surgical techniques have been developed and improved over the years. Delayed primary anastomosis is one of the most preferred choices for treatment of LGEA [4,20,21]. This method implies that the elongation of esophageal pouches benefits from the spontaneous growth potential of the esophagus and/ or the use of one of the mechanical traction techniques.

### ***Open and thoracoscopic approach***

Regarding the method for opening the thorax there are two possible approaches – open (thoracotomy) and thoracoscopic. In the recent past and probably still in some centers, thoracotomy is a preferred technique, regardless of the following stages of treatment, including the approximation of esophageal pouches [22]. The open technique is usually performed extrapleurally through a posterolateral thoracic wall via the right fourth intercostal space, below the inferior angle of scapula [7,21]. However, at a time of minimal invasive surgeries there are many reports of the efficient thoracoscopic management of LGEA [11,23–26]. The experts of the ERNICA Consensus Conference reported that thoracoscopy is a viable option, although only after suitable evaluation [4]. This recommendation may have been couched in this form because the laparoscopic approach is very challenging for the surgeon, and requires experience and specific competencies, such as a laparoscopic suturing. Nevertheless, the thoracoscopic approach has many advantages, both for the patient and the surgeon. Firstly, it requires only three incisions for inserting ports in the right pleural cavity (max. 5 mm port) which reduces scarring and, in consequence, the incidence of musculoskeletal deformations and chronic pain [10,27]. Researchers have also noted that a thoracoscopic repair shortens the total hospital length of stay and the time to first oral feed, and improves the recovery after surgery [23]. Moreover, a thoracoscopy enables better visualization and identification of esophageal pouches, as well as adjacent anatomical structures, and more precise tissue incisions. Thanks to this method blood loss and the risk of patient hypothermia are also minimized [20,27].

### ***External and internal traction sutures***

The gap between both esophageal ends in LGEA usually exceeds three intervertebral spaces, therefore an early primary anastomosis is not possible to perform. The time to restore the esophageal continuity is disputable [7]. Some researchers prefer to wait for the spontaneous growth of the esophagus, which has been reported to occur usually between 8 and 12 weeks of life [28]. Moreover, when the infant is fed through a gastrostomy a gastroesophageal reflux is considered as a factor which contributes to thickening of the lower esophageal pouch tissue and makes it more resistant to mechanical damage such as suturing [2]. However, several methods to stimulate an elongation of esophageal pouches and shorten the time to a delayed anastomosis have been developed. In the 1990s two approaches were proposed. First, the staged extrathoracic esophageal elongation technique, described by Kimura and second – the multistage tractional lengthening method, described by Foker [12,13]. Kimura's method

considered the elongation of the proximal pouch. It assumed forming subcutaneously an esophagostomy and moving it gradually down the anterior thoracic wall [8,10]. Foker proposed an extensive dissection of the proximal and distal esophagus, followed by placing traction sutures with their externalization through the chest wall and fixation [8,10]. In some cases, external traction needed to be preceded by the internal traction, during which the lumen of the esophageal pouches was not opened, the esophageal ends were placed into silicon sacks and sewed onto the prevertebral fascia.

Initially, the traction techniques were performed using the open approach. Van der Zee et al. undertook a successful attempt to elongate esophageal pouches thoracoscopically [29]. A major challenge for the surgeon while performing a thoracoscopic operation is suturing the esophagus. This is because the surgeon's efficient laparoscopic suturing technique is required [27]. There is also always a risk of tearing the esophagus when driving the needle and tying knots [26]. However, Bogusz et al. showed that the risk of perforation was higher in modifications of Foker's technique than when performing the multistage thoracoscopic surgery using the internal traction sutures [11]. This is a novel procedure which enables the elongation of the proximal and distal esophagus directly towards each other using sliding knots. Fixation on the thoracic wall is therefore not necessary [11,24,30]. Toczewski's et al. study conducted on an animal model (white Pekin Duck esophagus) showed that when using this technique pledget sutures or a double clip should be used, because the maximal force of traction without tissue disruption and the greatest elongation of esophageal pouches were then achieved [30]. Furthermore, this study showed that gradual tightening of the sliding knot in short periods (intermittent traction) enables tissue adaptation, reduces the risk of ruptures, and should be therefore recommended.

For ensuring early enteral feeding, optimal nutrition, and to promote stomach growth the experts recommend creating a gastrostomy prior to any surgical treatment of LGEA [4,21]. Moreover, as mentioned before, gastroesophageal reflux seems to have a positive impact on the distal esophagus making the walls thicker and more refractory [2]. In many cases a gastrostomy is created on the first or second day of life [2,21,25,31–34]. Although it precedes many delayed anastomosis surgeries, it seems that the use of the traction technique enables the omission of this stage [11,29]. This is possible because the mechanical elongation shortens the time between stages of surgical treatment. Furthermore, the possible complications connected with creating a gastrostomy are eliminated [11]. Regarding feeding, without the gastrostomy patients require parenteral nutrition. Some researchers perform a

laparoscopic gastropexy against the anterior abdominal wall to prevent the stomach from migrating and to fix it in the abdomen[29,31]. This procedure is not obligatory, though.

### ***Novel techniques***

Another innovative and effective technique in the treatment of LGEA is magnetic compression anastomosis (MCA)[14,32,33,35]. As with all the procedures mentioned above, this method also enables the preservation of the native esophagus [25] . It uses an electromagnetic field to perform the delayed anastomosis, avoiding dissection of the esophageal pouches which reduces the risk of injuring nerves, vessels and the trachea adjacent to the esophagus[33]. Zaritzky et al. suggested that this technique can be used both for elongation and anastomosis, but only in the cases in which the gap between the proximal and distal esophagus does not exceed 3 cm[14]. Although an anastomosis was achieved in all of Zaritzky's cases, almost all procedures were complicated by the anastomotic stenosis, requiring several dilations. Ellebaek et al. also reported the use of magnetic force to approximate the esophageal pouches [33]. However, in this case the growth of the proximal esophagus was stimulated by repeated gently mechanical force on the esophageal tube. The magnets were inserted after 2 months when the distance between esophageal pouches was reduced to 5 mm, achieving successful anastomosis. The diameter and the strength of the magnets have differed over the years. The most recent research reports a novel device using 8-mm-diameter convex-concave magnets[25]. The researchers highlight that MCA should be used only for an anastomosis and the thoracoscopic traction techniques should precede use of the magnets. They hypothesize that the longer the proximal and distal esophagus are, the more tension is reduced, and the less frequently anastomotic strictures will occur. Not only are the elongation procedures recommended but so also is the creation of a gastrostomy prior to MCA[25,32,33]. In addition to enteral feeding, it provides easy access for insertion of the lower magnet. The major advantages of MCA are avoidance of a thoracotomy and its long-term consequences, shorter surgery times - because of the thoracoscopic approach - and lower leakage rates[25,33,36]. Unfortunately, although effective in anastomosis and very promising, MCA needs to be improved in order to decrease the anastomotic stricture rate, the most frequent complication after performing this procedure.

The spontaneous growth and/or mechanical elongation techniques are not the only methods which have been described and developed to bring together the proximal and distal esophagus. A further promising method, which has the potential to shorten the time of delayed primary anastomosis or even enable early primary anastomosis, is the intramural botulinum type A toxin (BTX-A) injection. Several studies in animal models have been



reported, including a rat, piglet and rabbit model [37–40]. The results are auspicious. They showed that a BTX-A injection improves esophageal stretching features, resulting in a significant increase in elongation at maximal tension [37]. Moreover, BTX-A may have a positive effect on healing and contribute to a decrease in the frequency of anastomotic strictures [37,39]. Ellebeak et al. reported a successful attempt to use this method on a human and concluded that the BTX-A seems to have a similar effect on smooth muscles as on the striate muscles [15]. Nevertheless, further studies are essential in order to recommend this procedure as a stage of treatment for LGEA.

### ***Replacement techniques***

Even though there is no tissue or organ which can replace the functions of the esophagus in a satisfactory manner, the experts claim that replacement techniques are a viable option in LGEA treatment [4,21]. According to the study conducted by Stadil et al. in Finland and Denmark, replacement methods, especially gastric pull-up, were performed even more frequently than delayed primary anastomosis [41]. However, gastric, jejunal and colon transposition are challenging and connected with high morbidity and mortality rates; therefore it is recommended to perform them, in the order listed, only if it is impossible to preserve the native esophagus [4,11,19]. A detailed description of replacement methods is, however, beyond the scope of this article.

### ***Tissue engineering***

Probably the most future-oriented research on LGEA treatment is developing an esophageal substitute with tissue engineering techniques [16,42,43]. The approaches of using synthetic materials e.g., silicon combined with biological elements and absorbable structures did not however bring the expected results. The incidence of leakage and anastomotic strictures was high, and the esophageal functions could not be restored [16,44]. Nevertheless, the decellularized matrices seem to be promising. They can be derived naturally and then decellularized, preserving the extracellular matrix structure and composition and providing scaffolding for the growth of particular cells [16]. The studies on revascularization using proangiogenic factors and on the engineered matrix with absorbable stents preventing strictures are still ongoing. The goals which researchers have established include developing the circumferential tubular construct with growth potential prepared with autologous cells. These features will help to reduce the number of surgeries required and the need for a prolonged immunosuppressive treatment. Furthermore, the ideal substitute should have lasting qualities in a storage environment, so that the reconstruction surgeries could be performed in the first days of patient life using constructs prepared in advance [16].

Unfortunately, it will probably take years to develop the desired esophageal substitute, although the current achievements and future ideas are inspiring.

### ***Early and long-term outcomes, recommendations***

The most common complication after EA anastomosis is anastomotic strictures (AS)[3,17,45]. ESPGHAN-NASPGHAN in the Guidelines define AS as a narrowing at the level of the esophageal anastomosis and suggested that it should be considered clinically relevant only in symptomatic patients and those who are unable to achieve feeding milestones[46]. The panel recommends that the diagnosis of this complication can be made either by contrast study or endoscopically[46,47].

Many studies have been conducted to find and analyze potential risk factors of AS [9,17,48–50]. LGEA itself, as a type of EA, as in the described case, and anastomosis under tension are mentioned by most researchers [17,47,48,50] . Some of them also suggest that the higher number of sutures applied may cause this complication [9] . Individual studies reported that the incidence of AS may be higher when a thoracoscopic approach is performed [49,51] . However, a meta-analysis indicated no significant difference between an open and thoracoscopic technique in treatment of EA[23].

Prophylactic dilation of the esophagus remains one of the more discussed issues. The research conducted by Koivusalo et al indicated no difference in outcomes for the group of patients who underwent a routine stricture dilation versus the group who did not undergo a dilation intervention until the AS become symptomatic [52] . The ESPGHAN–NASPGHAN experts found no other evidence that the more invasive routine dilation method has more benefits than the selective dilations only when symptoms arise [46,47] . Therefore, the recommendation is to dilate the AS only when symptoms occur [53] . There is no evidence and/or consensus regarding the ideal dilation technique, or regarding the interval between dilation procedures [46] . The experts recommend the use of either balloon or semi-rigid (bougie) dilators [46,48] . Bougie dilators enable a surgeon to adjust appropriate force to the perceived resistance at the level of stenosis and are more cost-effective than balloon dilator, because they are reusable. The decision regarding the dilation method though should be dictated by the surgeon’s skills and preferences[47].

Potential adjuvant treatment is considered, especially in treatment of refractory strictures. Such strictures are defined as ‘inability to successfully remediate the anatomic problem to obtain age-appropriate feeding possibilities after a maximum of 5 dilation sessions with maximal 4-week intervals’ [53]. In such cases in children the ESGE/ESPGHAN suggest the topical application of mitomycin C following dilation. Other possible adjunctive therapy is

temporary stent placement [46,47,53]. However, no controlled studies have been conducted for these methods of AS treatment[46].

There are opinions amongst the experts that patients diagnosed with LGEA should be referred to the clinics specialized in treatment of this congenital defect and receive suitable expert evaluation [4,16,19]. Van der Zee et al. term these institutions as the Centers of Expertise (CoE) and define them as ‘pediatric surgical centers that are equipped and experienced in the treatment of patients with LGEA’ [19]. Lee et al. emphasizes that these clinics should additionally offer prenatal parental consultations and neonatal intensive care [16]. If an infant is diagnosed postnatally or a birth is not planned in the specialized center, it is still recommended that the infant be referred to the CoE, because an esophageal reconstruction is not performed right after the birth and the advantages outweigh the concerns e.g., transport issues[19]. Experience and proper equipment enable the use of the latest, novel surgical techniques, may shorten operating times, and prevent and treat possible complications.

Due to the accompanying abnormalities and the morbidity after surgical treatment patient with LGEA always requires intradisciplinary follow-up care. Multiple outpatient clinical and nutritional assessments are highly recommended [4]. Moreover, the experts recommend beginning screening endoscopic examinations already in adolescence[20]. Gastroesophageal reflux (GER) is a frequent complication after surgical treatment of LGEA. The incidence of GER hovers from 17% to 75% and is higher than in the general population [20,46]. The symptoms are prolonged and do not resolve over time. Gastroesophageal reflux disease (GERD) is a known risk factor for Barret’s esophagus – a precancerous condition which, untreated, can lead to esophageal cancer. Therefore, the experts emphasize the importance of screening esophagoscopy and endoscopic biopsies in the prophylaxis of Barret’s esophagus and cancer [20,53]. Baird et al. suggest that the frequency of endoscopic dilations should depend on esophageal lesions. The recommendation is to perform endoscopic examination and/or multistage biopsies every 5-10 years in patients without Barret’s esophagus, every 3 years in patients with Barret’s esophagus without dysplasia and every 6 months in patients with Barret’s esophagus with dysplasia[20].

Unfortunately, most of the foregoing recommendations are experts’ opinions or have a weak level of evidence. Therefore, further, preferably prospective, studies are necessary. Such research would have great value and would help to better understand and prevent the complications associated with surgical treatment of LGEA.

## **Conclusion**

Elongation techniques are effective in approximation of the proximal and distal esophagus. The future lies with tissue engineering and inventing an off-the-shelf esophageal substitute. Regardless of the surgical approach a common complication of surgical treatment is anastomotic stenosis which requires further surgical interventions. The centralization of treatment is recommended. Infants with LGEA should be referred to experienced and well-equipped clinics. After discharge from hospital interdisciplinary outpatient assessment and care is required. Screening endoscopic examination, usually combined with multistage biopsies, are recommended in adolescence and adulthood in the prophylaxis of Barret's esophagus and cancer. Further prospective studies are needed to determine the optimal mode of treatment and prevent complications associated with LGEA.

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### ***Authors' contribution***

Conceptualization, Z.K.; Investigation, Z.K., K.D., H.C-K., K.A., K.S.; Writing – Original Draft Preparation, Z.K.; Writing – Review & Editing, K.D., D.M., A.Ś., I.T., U.Ż., W.S. All authors have read and agreed with the published version of the manuscript.

### ***Conflict of interest***

The authors declare no conflict of interest regarding this article.

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